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Case Report

Parachute deformity of both atrioventricular valves with congenitally corrected transposition in an adult

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ABSTRACT

A 23-year-old female presented with mild exertional dyspnoea and palpitation since early childhood. By deploying 2D- and 3D echocardiography, she was detected to have situs solitus, atrioventricular and ventriculoarterial discordance with L-malposition of great vessels, valvular pulmonary stenosis, large secundum atrial septal defect, bicuspid aortic valve, right-sided aortic arch, and moderately severe mitral and tricuspid valve regurgitation. Typical parachute deformities of the morphologically mitral and tricuspid valves were observed. 3D echocardiography revealed a single papillary muscle in the morphologically left ventricle placed anteriorly and providing insertion to tendinous cords and only a moderator band with no other muscle bundles in the morphologically right ventricle placed posteriorly and providing attachment to two strings of cords. Considering the minimal symptoms, conservative treatment was pursued.

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1. Introduction

Unifocal insertion of the tensor apparatus of the atrioventricular valve has been called the parachute deformity by Jesse Edwards and his group [1]. The analogy to a parachute is suggested for its resemblance with the shape of the deformed valve. The leaflets resemble parachute of the chordae, its shrouds or strings, and the papillary muscle, the harness. The pathognomonic 'pear' shape of the mitral or tricuspid valve is seen in the four-chamber view, with the atrium forming the larger base of the pear and the leaflets the apex. Dominantly but not entirely focalized insertion of chords in the absence of a single muscle group has been called the parachute-like asymmetric valve which is probably much more common than is appreciated by imaging techniques. This report describes a

young woman with corrected transposition of great vessels in association with true parachute mitral and tricuspid valves.

2. Case report

A 23-year-old medium-built lady was examined for complaints of mild exertional dyspnoea and palpitation since early childhood. She had two successful pregnancies in the past and both children were reported to be normal. On examination, she was 164 cm tall, with a weight of 47 kg, blood pressure in the right arm 100/80 mmHg, and heart rate of 86 beats/min. There was no evidence of failure or cyanosis although pulse oximetry showed arterial saturation of 89%. Cardiovascular examination revealed a wide fixed split of second heart sound with A₂ louder than P₂ and a grade 4/6 ejection systolic murmur in the left upper

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parasternal area. The 12-lead electrocardiogram showed north-west axis ($+150^\circ$), right atrial enlargement and prominent R wave in V_1 only. The chest skiagram showed mild enlargement of cardiac silhouette with aortic shadow on the right side. 2D echocardiography showed situs solitus, atrio-ventricular discordance, L-loop of the ventricles, ventriculo-arterial discordance, right-sided aortic arch, valvular pulmonary stenosis, bicuspid aortic valve, large secundum atrial septal defect with predominant left to right shunt and moderately severe mitral and tricuspid regurgitations (Figs. 1-2). There was a single papillary muscle in the morphologically left ventricle inserted high up in the anterolateral wall with normal length and thickness of the tendinous chords, and mitral regurgitation occurred because of symmetrical tethering of the leaflets (Figs. 2-3). Morphologically right ventricle showed increased wall thickness and two strings of chords directly attached in the middle of the moderator band (MB, Figs. 4-5). No intervention was planned in view of mildly symptomatic status and the patient's reluctance.

3. Discussion

Congenital anomalies of the atrio-ventricular valves represent a wide spectrum of lesions that are often associated with other

congenital heart defects. The malformation of an atrioventricular valve in which the chordal apparatus is inserted into a single papillary muscle or a muscle group is called parachute deformity.¹ The "parachute mitral valve" has the usual two mitral valvular leaflets and commissures, but the chordae, instead of diverging to insert into two papillary muscles, converge into one major papillary muscle. In "true" parachute mitral valve, mitral valve chordae insert into a solitary papillary muscle. Initially atrioventricular valves with unifocal attachment of chords were loosely called "parachute valves," independent of the number of papillary muscles. Because of the focalized attachment of chords to one of the papillary muscles in the ventricles, some called this malformation "parachute-like asymmetric mitral valve or parachute-like tricuspid valve".² In parachute-like mitral valve,³ chordal attachments are more common to the posteromedial papillary muscle (73%) than the anterolateral papillary muscle (27%). Shortly after its description involving the mitral tensor apparatus, it was observed that often, the anomaly was associated with left-sided obstructive lesions and the syndrome was called the Shone complex.⁴ Parachute tricuspid valve is a late discovery⁵ with a single case each reported with tetralogy of Fallot,⁶ double-outlet right ventricle,⁷ two cases with atrial septal defect^{8,9} and three in isolation.¹⁰⁻¹² Most of these cases were not symptomatic due to parachute tricuspid valve.

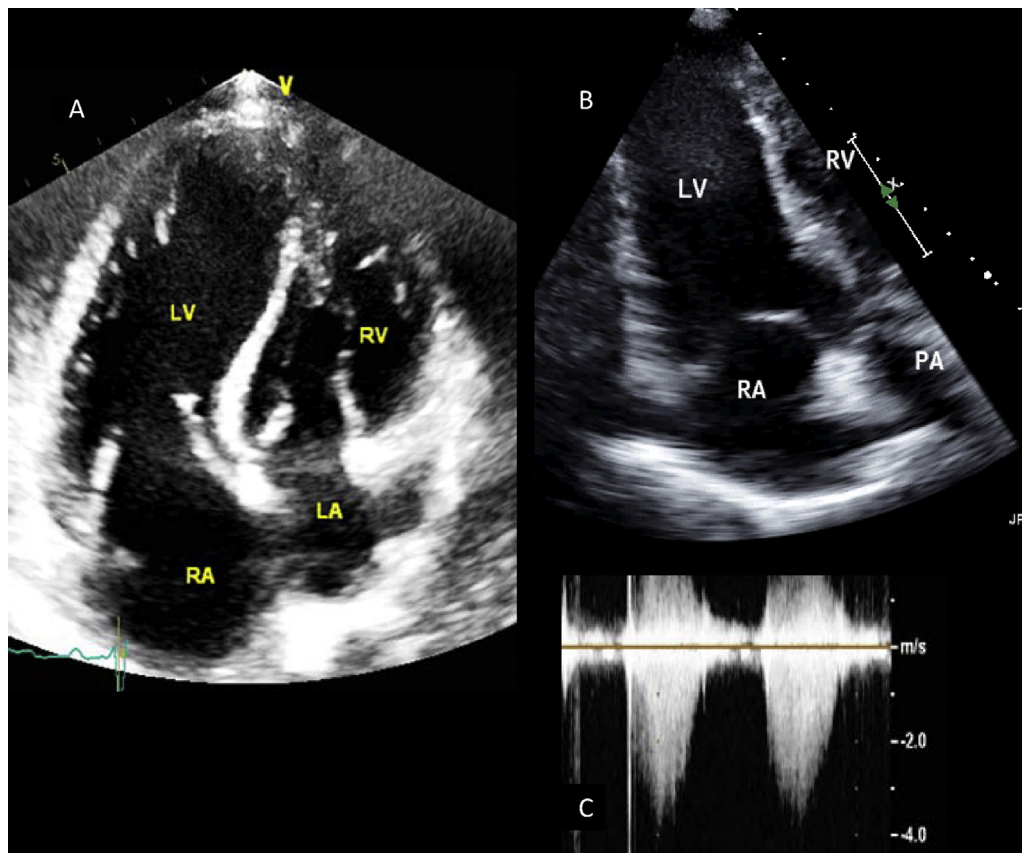


Fig. 1 – (A) Composite picture showing atrio-ventricular and ventriculoarterial discordance. The main pulmonary artery **(B)** originates from the morphologically left ventricle and shows valvular stenosis with a peak gradient of 70 mmHg across the pulmonary valve **(C)**. Note the parachute-like appearance of the morphologically tricuspid valve **(A)**. Atrial septal defect is clearly visible.

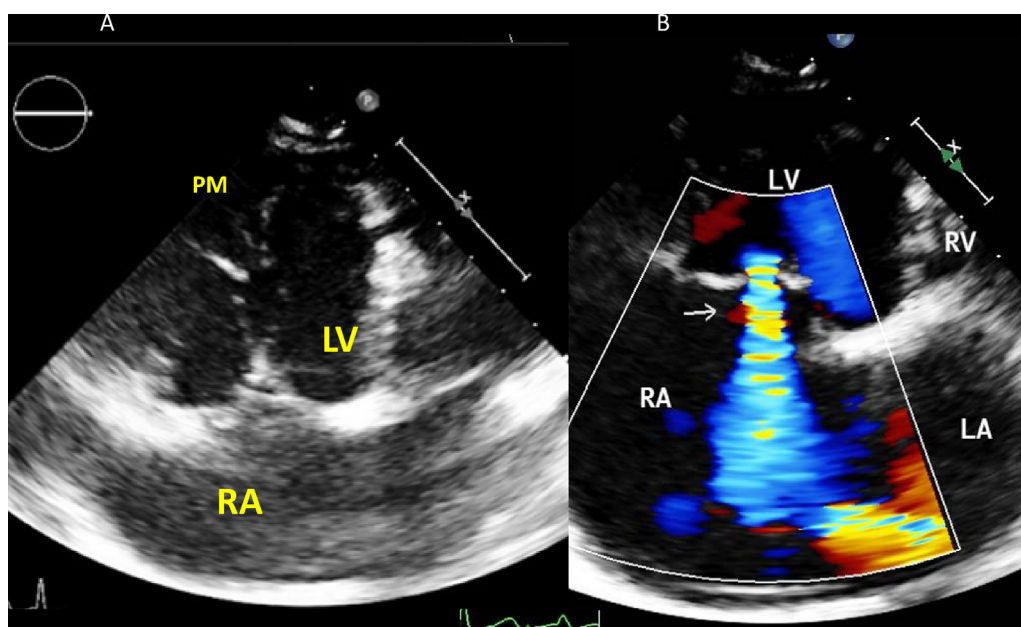


Fig. 2 – Abnormally high insertion of the single papillary muscle (PM) in the morphologically left ventricle with unifocal attachment of chords (left panel). The right panel shows significant mitral regurgitation on color flow mapping.

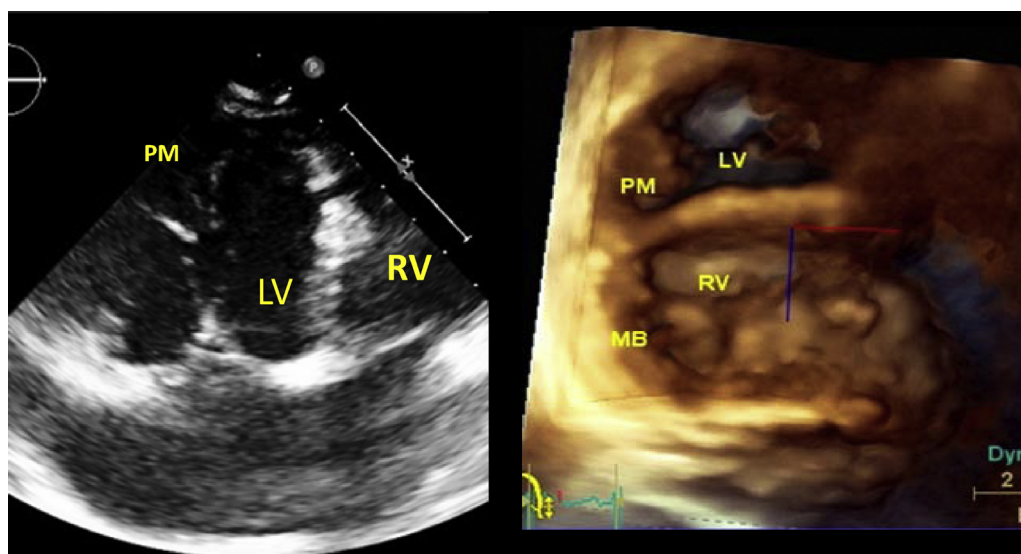


Fig. 3 – Demonstration of a single papillary muscle by 2D and 3D (right panel) echocardiography in the morphologically left ventricle (LV). PM – papillary muscle, MB – moderator band.

Congenitally corrected transposition is a rare cardiac malformation characterized by the combination of discordant atrioventricular and ventriculo-arterial connections, usually accompanied by other cardiovascular malformations. Associated malformations may include interventricular communications, obstructions of the outlet from the morphologically left ventricle, and anomalies of the tricuspid valve. In corrected transposition of great vessels, congenital mitral valve abnormalities have been described in 55% of the cases; of which <10% have parachute-like mitral valves; tricuspid abnormalities have been reported in 86% cases but none with parachute

tricuspid valve.¹³ Incidentally, first description of the parachute mitral valve was reported in patients with corrected transposition.¹ In a report of 52 patients with parachute mitral valve, Tandon et al.¹⁴ reported two cases with congenitally corrected transposition and none of the cases had associated parachute tricuspid valve. Parachute deformity of both the atrioventricular valves has been reported so far only in one case of ventricular septal defect.¹⁵

This patient had parachute deformities of both the atrioventricular valves causing valvular regurgitation in the setting of congenitally corrected transposition of great vessels (double

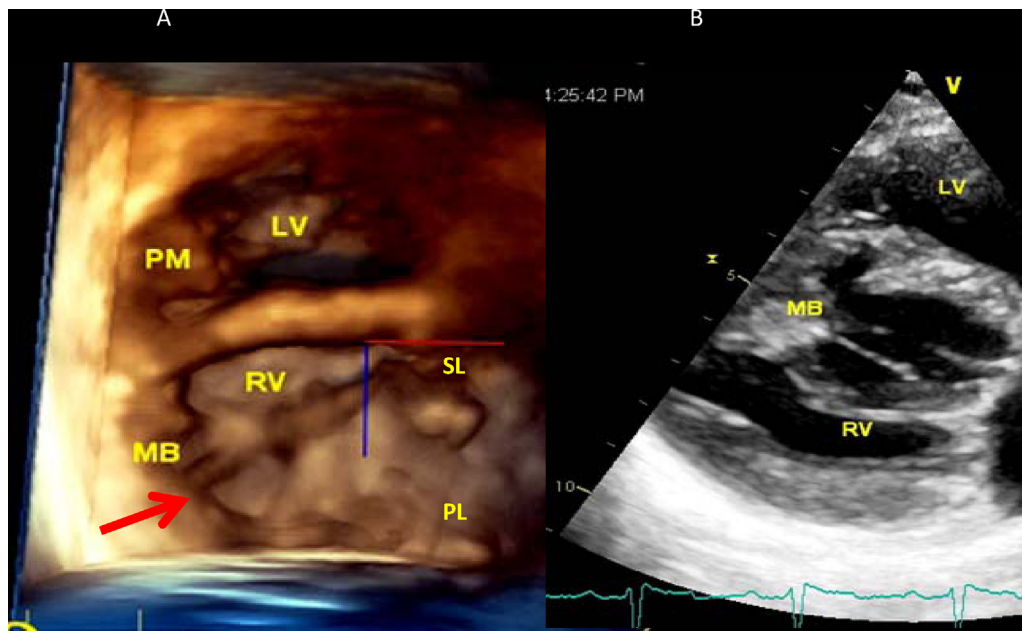


Fig. 4 – Unifocal insertion of the chords in the moderator band during diastole (left panel) and during systole (right panel). Posteriorly located morphologically right ventricle.

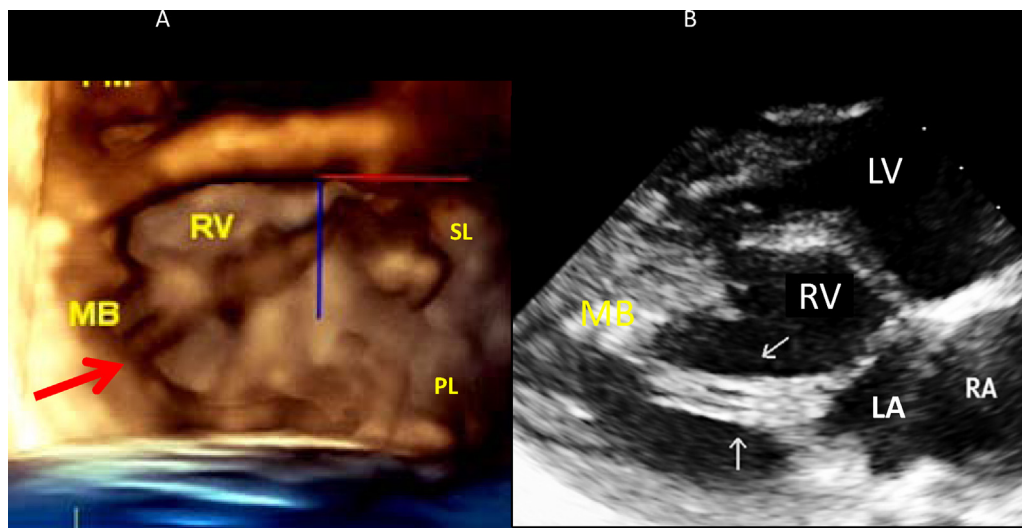


Fig. 5 – Focused views of the morphologically right ventricle showing no muscle groups other than the moderator band. The two strings of the chords in diastole (left panel) form a “parachute”.

discordance). 3D echocardiography was of incremental value in delineating the papillary muscles and the unifocal attachment of chords. Both valves assumed pear-shaped canopy appearance during diastole and two distinct strings of chords which crowded in the center of the ventricular cavities during systole. Extensive literature search showed one such case along with ventricular septal defect reported previously.¹⁵ There are reports of parachute mitral valve in patients with corrected transposition^{1,13,14} but not of parachute tricuspid valve. More often, there is Ebstein anomaly of the left-sided atrio-ventricular valve which can cause valvular regurgitation

in corrected transposition. Marino et al.³ also reported that most patients with parachute mitral valve did not have mitral valve regurgitation at initial echocardiogram, and only a small number of patients had progression to hemodynamically significant mitral valve regurgitation. It is unknown how long our patient has mitral valve regurgitation. However, according to the report, patients with parachute valve may be at risk for progressive mitral regurgitation and should be followed up. Occasionally, severe mitral regurgitation may be the only presentation of the parachute mitral valve.¹⁶ Presence of bicuspid aortic valve and valvular pulmonary stenosis may

indicate incomplete Shone complex on both sides. Parachute deformity may be benign in a number of cases and detected incidentally. However our patient had significant atrio-ventricular valve regurgitation which may become symptomatic in future.

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